In the Specification:

Please amend paragraph [8] as follows:

8. The present invention <u>is</u> ~~if~~ based, in part, on the discovery that NB-DNJ administered to mice together with glucocerebrosidase (the major therapy for Gaucher Type I patients) unexpectedly does not compromise the activity of glucocerebrosidase, and further, provides an augmentation of enzyme activity over time due to a protective effect of NB-DNJ on the enzyme. This result is surprising as the efficacy of the enzyme would be expected to be compromised in the presence of NB-DNJ, as NB-DNJ is a weak inhibitor of glucocerebrosidase (IC_{50} = 0.52 mM). It has further been discovered that the co-administration of NB-DNJ with bone marrow transplantation to provide enzyme augmentation to increase the rate of neuronal glycolipid degradation provides an unexpected synergistic effect.

Please amend paragraph [27] as follows:

27. In the context of the present invention, the term "**inhibitor**" is intended to include inhibitors which inhibit glucosylceramide synthesis. It includes molecules such as N-butyldeoxynojirimycin, N-butyldeoxygalactonojirimycin, N-nonyldeoxynojirimycin and other imino sugar-structured inhibitors of glucosylceramide synthesis. It also <u>includes</u> ~~includses~~ other inhibitors of glycolipid synthesis, especially glucosylceramide synthesis, including agents such as 1-phenyl-2-decanoylamino-3-morpholino-1-propanol (PDMP), D-threo-1-phenyl-2-decanoylamino-3-morpholino-1-propanol and structurally related analogues thereof. Further, inhibition can also be achieved by the use of genetic approaches, based on the introduction of nucleic acid coding for proteins or peptides capable of inhibiting glycolipid synthesis or antisense sequences or catalytic RNA capable of interfering with the expression of enzymes responsible for glycolipid and especially glucosylceramide synthesis (e.g. glucosylceramide synthase). A combination of any of the above inhibitors can be used.

Please amend paragraph [29] as follows:

29. The term "**substantially pure**," when referring to a polypeptide, means a

polypeptide that is at least 60%, by weight, free from the proteins and naturally-occurring organic molecules with which it is naturally associated. A substantially pure glucosylceramide synthesis inhibitor is at least 75%, more preferably at least 90%, and most preferably at least 99%, by weight, glucosylceramide synthesis inhibitor. A substantially pure glucosylceramide ~~glucosylceramie~~ synthesis inhibitor such as N-butyldeoxynojirimycin (NB-DNJ), can be obtained, for example, by chemical synthesis or by isolation from natural sources. Purity can be measured by any appropriate method, e.g., column chromatography, polyacrylamide gel electrophoresis, or HPLC analysis.

Please amend paragraph [34] as follows:

34. The galactose analogue of NB-DNJ, N-butyldeoxygalactonojirimycin (NB-DGJ), is known to inhibit GSL synthesis in vitro as effectively as NB-DNJ, but is more specific in that it does not inhibit α-glucosidase I and II or β-glucocerebrosidase (Platt et al, (1994) J Biol Chem 269(43): 27108-14). It is known that only approximately 10% of the serum level of NB-DNJ is present in the cerebrospinal fluid. Accordingly, high systemic doses of NB-DNJ may have to be administered in order to achieve therapeutic levels in the CNS, and may have to be administered for the duration of a patient's ~~patients~~ life. High concentrations of NB-DNJ in humans causes diarrhoea and in mice it causes weight loss and reduces the size of lymphoid organs. Thus, it would be advantageous to have an inhibitor of glucosylceramide synthesis which does not have these disadvantages of NB-DNJ.